Alerts, Notices, and Case Reports

An Unusual Cause of Dysphagia

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MYASTHENIA GRAVIS has an annual incidence of 3 to 4 per million.[1] The diagnosis is often missed because it is rare and because its symptoms characteristically fluctuate. We report a case of myasthenia gravis in a patient who presented with dysphagia. The patient had been seen by several specialists who had performed a multitude of tests without reaching the correct diagnosis. Symptoms of dysphagia differ according to the cause. Both neuromuscular and esophageal causes must be considered in the evaluation of dysphagia.

Report of a Case

The patient, a 37-year-old man, was admitted to University Medical Center of Southern Nevada (Las Vegas) after three months of progressive dysphagia and an 11-kg (25-lb) weight loss. Six years before admission, the patient had suffered from nasal regurgitation of liquids, with tracheal aspiration and coughing, and diplopia when driving late at night. The nasal symptoms and aspiration resolved spontaneously within three weeks, but the diplopia persisted. Three months before admission, he began having difficulty swallowing solids. The dysphagia worsened, extending during the next two months to liquids. The patient again had nasal regurgitation of liquids, along with coughing from tracheal aspiration. His symptoms were noticeably worse in the afternoon and evening. He saw a gastroenterologist, a surgeon, and an internist. A barium swallow, esophagogastroduodenoscopy, computed tomographic scans of the abdomen and chest, esophageal motility studies, and esophageal manometry revealed a right middle lobe infiltrate and mild gastritis. At the time of admission, he was unable to swallow solids or liquids, but did not have odynophagia. In addition to dysphagia and diplopia, the patient complained of easy fatigability of his middle fingers, which he used extensively in his job as a newspaper carrier. His medical history was notable for head trauma seven years earlier, resulting in the loss of all upper teeth. He recovered fully and had no neurologic deficits.

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On physical examination the patient was thin but in no acute distress. Mild bilateral ptosis was present, but vision and extraocular movements were grossly intact. He had symmetrical weakness of the facial muscles. His gag reflex was weak, and he was unable to swallow. He could extend his tongue only 1 cm beyond his lips. Speech was markedly nasal and slurred. The rest of the physical findings were within normal limits. Blood chemistry values and blood counts were unremarkable except for a serum cholesterol level of 2.8 mmol per liter (110 mg per dl). Following the intravenous administration of 10 mg of edrophonium chloride (Tensilon), the patient could swallow and speak clearly. His ptosis resolved, and he regained full strength in the muscles of his face and tongue. A serum acetylcholine-receptor antibody titer was later found to be elevated at 12.5 ng per ml. Treatment with pyridostigmine bromide, prednisone, and intravenous immune globulin was begun, resulting in rapid but incomplete improvement. A few weeks later the patient underwent a thymectomy. There were no complications, and no thymoma was found. Prednisone therapy was discontinued, and the patient was maintained on a regimen of pyridostigmine after the operation.

Discussion

In retrospect, our patient had a fairly classic case of myasthenia gravis. The symptom constellation of dysphagia, diplopia, and facial muscle weakness, worsening late in the day, is typical of this disorder.[2] Obtaining a history of nighttime diplopia and fluctuating weakness requires specific, direct questions. A high level of suspicion for myasthenia gravis is necessary. Nighttime examination of patients may be useful. Because the symptoms of the disorder usually fluctuate, the results of a daytime examination may be completely normal. Dysphagia is the chief complaint in only 6% of patients with myasthenia gravis,[3] although 28% exhibit bulbar symptoms—dysphagia or dysarthria—at its onset.[2] Ocular symptoms—diplopia and ptosis—are the chief complaint in 53% of patients with myasthenia gravis. Weakness in the face, neck, trunk, and limbs may also occur; in general, muscular weakness has a highly variable pattern in patients with this disorder.[3]

The patient's major symptom was dysphagia, which, as mentioned earlier, is the main symptom in a minority of patients with myasthenia gravis. Therefore, the evaluation before admission to hospital focused on this symptom. The exact nature of dysphagia can vary depending on the disease that causes it. Dysphagia falls into two major categories: oropharyngeal and esophageal.

In oropharyngeal dysphagia, patients have difficulty with the initial phase of swallowing, that is, transferring a bolus from the mouth to the upper esophagus. The differential diagnosis of oropharyngeal dysphagia includes neurologic disorders (cerebrovascular accident, multiple

sclerosis) and motor unit disorders (amyotrophic lateral sclerosis, progressive bulbar palsy, muscular dystrophy, polymyositis or dermatomyositis, and myasthenia gravis). Symptoms of oropharyngeal dysphagia do not differ greatly in these various diseases. Dysphagia occurs with liquids and solids, and multiple attempts may be necessary to swallow successfully. Nasal and oral regurgitation often occur immediately after a swallowing attempt, sometimes with a forceful spraying of the mouth contents. Tracheal aspiration is common, possibly leading to pneumonia. In severe cases, patients may not even be able to swallow their own saliva, and malnutrition and weight loss may result.[4]

In esophageal dysphagia, the swallowing act is initiated normally, but a bolus of food fails to progress into the stomach. Patients typically describe a feeling of food sticking during swallowing. This sensation is generally in the esophagus but may be referred proximally to the neck, even with a distal esophageal lesion.[5] The differential diagnosis of esophageal dysphagia includes obstructions (rings and webs, benign strictures, and cancers) and motility problems (achalasia, scleroderma). Generally, obstructive disorders (unless severe) cause dysphagia for solids only, whereas motility disorders cause dysphagia for both liquids and solids. Heartburn is typical of scleroderma and strictures. Cancers cause a rapid progression of symptoms, and rings and webs usually cause intermittent dysphagia. Oral regurgitation, but not nasal regurgitation, may occur with esophageal dysphagia, generally hours after swallowing.[4]

Careful questioning can help to determine the cause of dysphagia. If the history suggests myasthenia gravis, an edrophonium test should be done. Although no precise figures have been published, sensitivity is estimated to be about 86% in the ocular form of the disease and 95% in generalized myasthenia—not limited to ocular symptoms. Specificity for the edrophonium test is not clear-cut, but a number of diseases are known to produce false-positive results, including amyotrophic lateral sclerosis and the Guillain-Barré syndrome. A serum acetylcholine-receptor antibody titer is the appropriate second-line confirmatory test, because specificity is high. Sensitivities of 64% (ocular) and 89% (generalized) have been reported.[6] These two tests are accurate and inexpensive enough that they should be done in almost every case of suspected myasthenia gravis.[6,7] Electromyographic techniques (conventional or single-fiber) can also be used to confirm the diagnosis. These techniques have a high specificity, and single-fiber electromyography is fairly sensitive but not widely available. Therapy includes both medical and surgical modalities, and established guidelines are available.[8-10]

In summary, myasthenia gravis is an often-overlooked cause of dysphagia. The differential diagnosis of dysphagia includes oropharyngeal and esophageal causes. When the history suggests oropharyngeal dysphagia, neurologic causes, including myasthenia gravis, must be carefully considered. A directed neurologic history and physical examination, possibly including a nighttime examination, is necessary and sufficient to diagnose most cases of this disorder.

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Nontraumatic Splenic Hematoma Related to Cocaine Abuse

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SPLENIC HEMATOMA and rupture are most commonly seen after blunt abdominal trauma. In rare instances splenic hematomas can occur without trauma, usually in patients with splenomegaly or underlying hematologic disorders.[1,2] This is the first report of a case of an otherwise healthy man in whom a splenic hematoma developed shortly after he used cocaine intranasally.

Report of a Case

The patient, a 40-year-old man who habitually used cocaine, presented to a local emergency department with left upper quadrant abdominal pain radiating to his left shoulder. Seven hours previously, he had "snorted" an unknown quantity of cocaine. He was working on his automobile when the pain began and became progressively severe. In the emergency department, his anterior chest wall was sensitive, but his lungs were clear, heart regular, and an abdominal examination revealed good bowel sounds. His abdominal wall was soft, without

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